

Pseudo-aplasia of the Right Pulmonary Artery Associated with Right-sided Aortic Arch

W. PORSTMANN, R. A. EL-SALLAB*, AND H. DAVID

*From the Cardiovascular Diagnostic Department and the Institute of Pathology, Faculty of Medicine (Charité),
Humboldt University, Berlin*

Congenital absence and hypoplasia of one pulmonary artery are anomalies now diagnosed with increasing frequency as the result of improvements in diagnosis during life. From 1850 to 1952 only 9 cases of congenital absence of one pulmonary artery had been reported (Madoff, Gaensler, and Strieder, 1952). In all these, the diagnosis was made at necropsy or thoracotomy. Christeller (1916) collected 34 cases of hypoplasia or aplasia of one pulmonary artery reported between 1816 and 1905, but he did not record the affected side. In 1960, Kröker found 140 cases reported between 1916 and 1960. In this material, the right side was involved in 17, the left side in 38, and in the remaining cases the affected side was not mentioned. Kröker made the diagnosis in his own cases by observing the difference in vascularity of the two lungs on plain radiographs and tomograms. In recent years, the wider use of angiocardiology has led to these anomalies being diagnosed more frequently. In the majority, additional congenital abnormalities are present: Fallot's tetralogy is a common associated anomaly; and the left pulmonary artery is more often affected than the right (Nadas *et al.*, 1953; McKim and Wigglesworth, 1954; Emanuel and Pattinson, 1956; Smart and Pattinson, 1956; Elder *et al.*, 1958; Sherrick, Kincaid, and DuShane, 1962; Pool, Vogel, and Blount, 1962; Bock *et al.*, 1963; Oakley, Glick, and McCredie, 1963; Oram, Pattinson, and Davies, 1964).

While valuable information can be provided by plain radiographs and tomograms of the lungs (Stecken, 1964; Richter, 1963), angiocardiology is still necessary to demonstrate the anatomy, especially in cases associated with cyanotic congenital heart disease.

Received May 17, 1966.

* Present address: Radiology Department, Faculty of Medicine, Alexandria University, Egypt, U.A.R.

Recently, Rockoff and Gilbert (1965) stated that angiocardiology could lead to a misinterpretation of the anatomy. They described the case of a 6-year-old cyanotic girl, with apparent atresia of the infundibulum of the right ventricle shown by angiocardiology. At catheterization and operation, however, the infundibulum was found to be stenosed only, and this stenosis was resected.

The purpose of this paper is to report another anomaly that may lead to the erroneous radiological diagnosis of aplasia of one pulmonary artery following plain films and angiocardiology. Our investigations in three cases are described.

CASE REPORTS

Case 1. This man, aged 26 years, had severe cyanosis from birth, and was found to have a harsh systolic murmur over the third left intercostal space. The plain films showed a globular heart shadow and a right-sided aortic arch. The left lung showed a normal vascular pattern, whereas the right lung showed an irregular network of vessels typical of a massive broncho-pulmonary collateral circulation. There was also prominent rib notching on the right side (Fig. 1). At cardiac catheterization via the right basilic vein, the left pulmonary artery was entered, and the typical findings of obstruction to the outflow tract of the right ventricle were recorded. At angiocardiology a few days later, the catheter passed up the left basilic vein, entered a persistent left superior vena and the left ventricle via an interatrial septal defect. The contrast medium was injected into the left ventricle, and passed through a ventricular septal defect, demonstrating stenosis of the infundibulum of the right ventricle and pulmonary valve stenosis. The aortic root partially arose from the right ventricle. A striking feature was the absence of filling of the right pulmonary artery and the prominent broncho-pulmonary circulation on that side (Fig. 2 and 3). A diagnosis of pentalogy of Fallot was made, with right-sided aortic arch, persistent left superior vena cava, and aplasia of the right pulmonary artery. It was

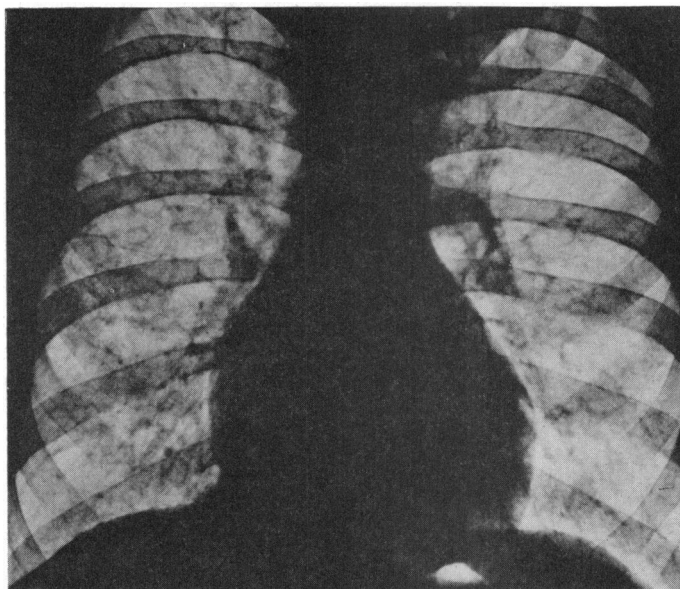


FIG. 1.—Case 1. Plain film showing globular-shaped heart with right-sided aortic arch, abnormal pattern of vessels in the right lung, slightly diminished vascularity of the left lung, and rib notching on the right side due to hypertrophied collateral systemic circulation.

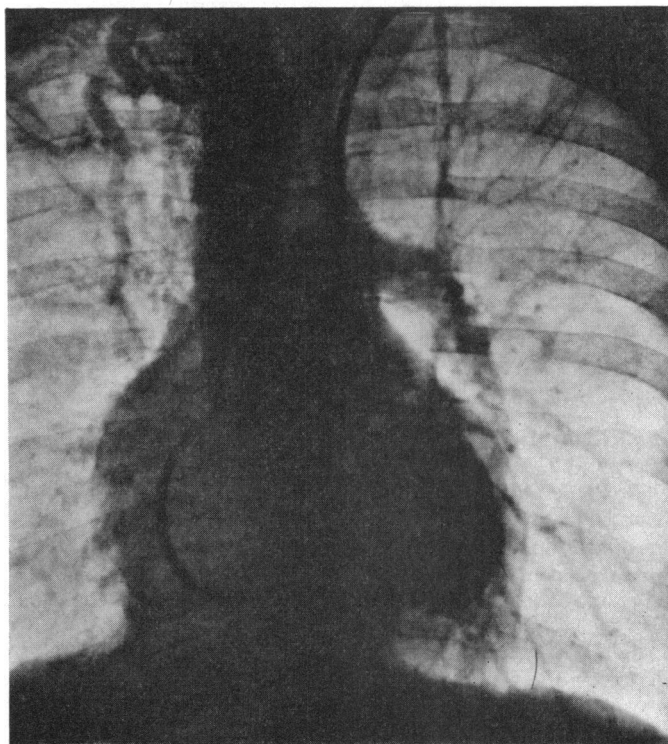


FIG. 2.—Case 1. Angiocardiogram. Left ventricular injection with filling of the ascending aorta and right-sided aortic arch. The main pulmonary artery and left pulmonary artery filled via the ventricular septal defect, but no filling of the right pulmonary artery was seen throughout the examination. Hypertrophied collateral systemic circulation on the right.

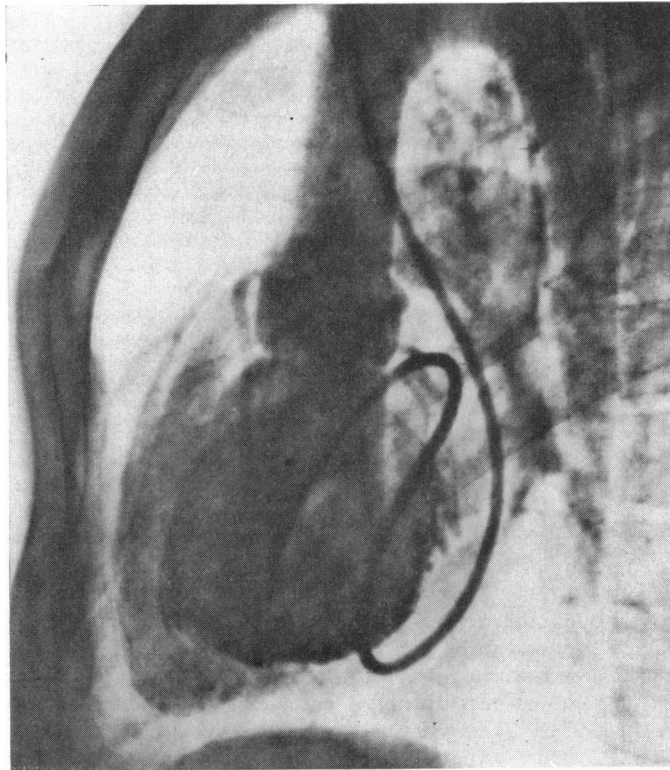


FIG. 3.—Case 1. Angiocardiogram, showing the right ventricle filling via the ventricular septal defect. There is stenosis of the infundibulum of the right ventricle and pulmonary valve stenosis. The latter is hidden. Enlarged bronchial arteries are filled.

decided not to attempt a surgical total correction of the defects in view of the presence of aplasia of the right pulmonary artery. Three years later the patient was readmitted to hospital in right ventricular failure, and died a few days later. At necropsy, the clinical and radiological findings were confirmed with one important exception. The right pulmonary artery was in fact patent and not stenosed, though its calibre was slightly less than that of the left pulmonary artery. The ascending aorta was a little dilated, and compressed the proximal part of the right pulmonary artery where it crossed the latter in the mid-line. The findings at necropsy are illustrated in Fig. 4.

Case 2. A man of 27 years, who had had severe cyanosis from birth, with a clinical diagnosis of Fallot's tetralogy, was referred for investigation. At angiocardiography, contrast medium, injected into the right ventricle, passed into the ascending aorta and right-sided aortic arch, and into the left pulmonary artery, but not into the right pulmonary artery at this time. The collateral broncho-pulmonary circulation on the right was faintly outlined (Fig. 5A). One and a half seconds later, the right pulmonary artery filled but was diminished in size, and there was also filling of the collateral circulation in the right upper lobe (Fig. 5B).

Case 3. A girl aged 3 years with cyanosis was referred with a clinical diagnosis of Fallot's tetralogy. At angiocardiography with injection into the right ventricle, the appearances were similar to those described in Case 2, except for the absence of a collateral circulation (Fig. 6A). One second later, there was early filling of the right pulmonary artery and increased filling of the left pulmonary artery and its minor branches. No collateral circulation was demonstrated (Fig. 6B).

DISCUSSION

Aplasia of one pulmonary artery as an isolated anomaly is of little clinical importance, and in association with congenital heart disease it is but a minor complication of more serious defects.

Angiocardiography is generally believed to be the most accurate method of demonstrating aplasia of one pulmonary artery before operation. In our first case, however, angiocardiography led to the mistaken diagnosis of aplasia of the right pulmonary artery which was shown to be patent and normally formed at necropsy. The secondary hypertrophy of the collateral broncho-pulmonary circulation on the right side supports the view that there was little

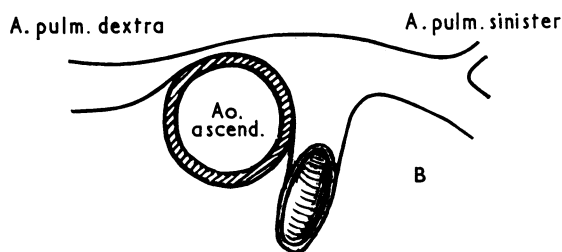
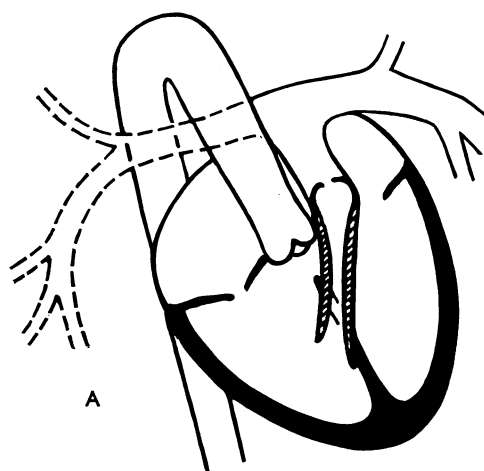


FIG. 4.—Case 1. Diagram of the pathological specimen. (A) Frontal view showing the transposed root of the aorta, the ascending aorta, crossing the proximal part of the right pulmonary artery and the right-sided aortic arch. (B) Transverse section seen from above showing compression of the right pulmonary artery by the ascending aorta.

or no blood flow through the right pulmonary artery during life. The angiocardigraphic appearances could be explained by extrinsic pressure on the proximal part of the right pulmonary artery. At necropsy this compression was found to be due to the dilated and partly transposed ascending aorta and was sufficiently severe to have almost completely occluded the blood flow through the right pulmonary artery during life.

The findings in the other two cases reported here seem to confirm this concept. Both were cases of Fallot's tetralogy with right-sided aortic arch, and in both angiocardiology showed the left pulmonary artery filling well, at least one second before the right pulmonary artery filled. The ascending aorta crossed the proximal part of the right pulmonary artery, and this artery was much smaller than the left pulmonary artery in both cases. In

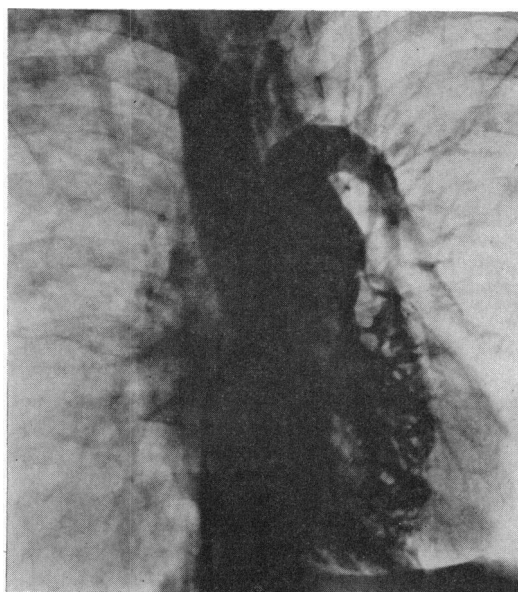


FIG. 5A.—Case 2. Angiogram. Contrast medium passes from the right ventricle into the ascending aorta and right-sided aortic arch and into the main and left pulmonary artery. The right pulmonary artery has not filled. There is faint filling of the collateral circulation on the right side.

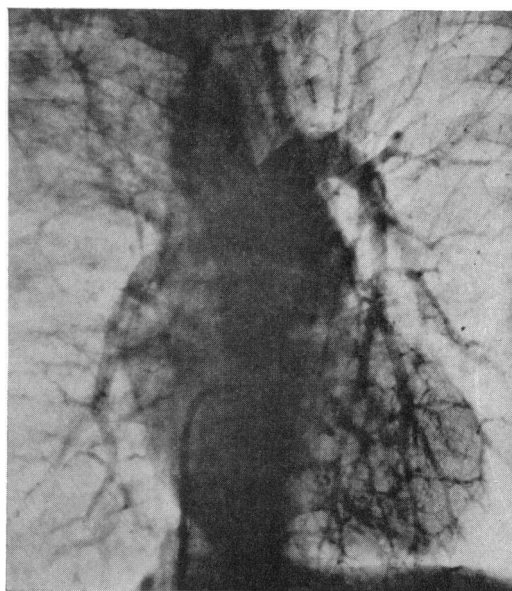


FIG. 5B.—The same case, $1\frac{1}{2}$ seconds later. The small right pulmonary artery has now filled and also the collateral circulation on the right side.

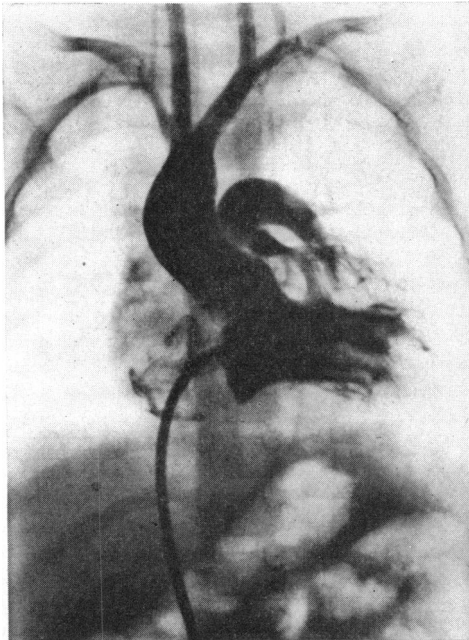


FIG. 6A.—Case 3. Angiocardiogram, with right ventricular injection. The appearances are similar to Case 2 except for the absence of a collateral circulation.

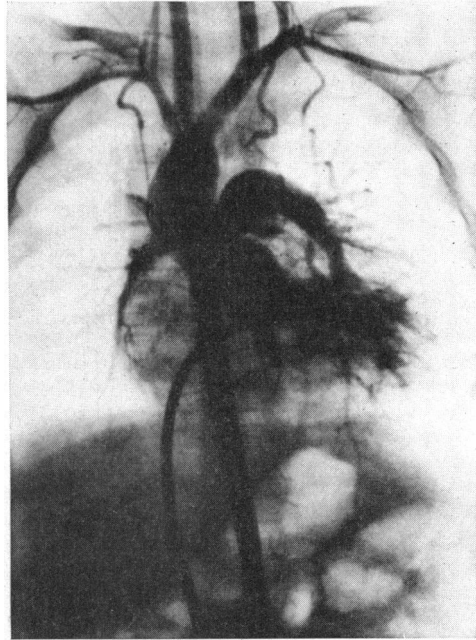


FIG. 6B.—The same case—1 second later. The small pulmonary artery has now filled but not as well as the left pulmonary artery.

Case 2, a prominent broncho-pulmonary circulation is shown on the right side in addition to the filling of the right pulmonary artery. This picture of varying degrees of reduced blood flow through the right pulmonary artery, associated with a right-sided aortic arch, may perhaps constitute a syndrome which, to our knowledge, has not been previously reported. Recognition of this syndrome may be important as it was in our first case, where operation had been ruled out in the mistaken belief that the right pulmonary artery was aplastic. Differentiation between extrinsic compression of a normally formed right pulmonary artery and aplasia of the artery must be made before the correct treatment can be selected. We suggest that in cases of Fallot's tetralogy with right-sided aortic arch and apparent aplasia of the right pulmonary artery, catheterization and selective angiography of the right pulmonary artery should be attempted. It may then be possible to differentiate between functional occlusion and true aplasia.

SUMMARY

A case of Fallot's tetralogy is reported in which a diagnosis of aplasia of the right pulmonary artery was made after study of the plain films and angiocardiology. There was a hypertrophied col-

lateral circulation to the right lung. At necropsy, a normally formed pulmonary artery was found. The radiological appearances suggestive of aplasia were produced by extrinsic pressure on the proximal part of the right pulmonary artery by the ascending aorta, the arch of which was on the right side. Two other cases with similar features but with delayed filling of the right pulmonary artery are described. This picture may constitute a functional syndrome.

REFERENCES

- Bock, K., Richter, H., Trenckmann, H., and Herbst, M. (1963). Die Aplasie einer Lungenarterie. *Fortschr. Röntgenstr.*, **98**, 419.
- Christeller, E. (1916). Funktionelles und Anatomisches bei der angeborenen Verengung und dem angeborenen Verschluss der Lungenarterie. *Virchows Arch. path. Anat.*, **223**, 40.
- Elder, J. C., Brofman, B. L., Kohn, P. M., and Charms, B. L. (1958). Unilateral pulmonary artery absence or hypoplasia. *Circulation*, **17**, 557.
- Emanuel, R. W., and Pattinson, J. N. (1956). Absence of the left pulmonary artery in Fallot's tetralogy. *Brit. Heart J.*, **18**, 289.
- Kröker, P. (1960). Zur Frage der sogenannten progressiven Lungendystrophie. *Fortschr. Röntgenstr.*, **93**, 1.
- McKim, J. S., and Wiglesworth, F. W. (1954). Absence of the left pulmonary artery; a report of 6 cases with autopsy findings in three. *Amer. Heart J.*, **47**, 845.

- Madoff, I. M., Gaensler, E. A., and Strieder, J. W. (1952). Congenital absence of the right pulmonary artery. Diagnosis by angiocardiology, with cardiorespiratory studies. *New Engl. J. Med.*, **247**, 149.
- Nadas, A. S., Rosenbaum, H. D., Wittenborg, M. H., and Rudolph, A. M. (1953). Tetralogy of Fallot with unilateral pulmonary atresia; a clinically diagnosable and surgically significant variant. *Circulation*, **8**, 328.
- Oakley, C., Glick, G., and McCredie, R. M. (1963). Congenital absence of a pulmonary artery. Report of a case, with special reference to the bronchial circulation and review of the literature. *Amer. J. Med.*, **34**, 264.
- Oram, S., Pattinson, N., and Davies, P. (1964). Post-valvular stenosis of the pulmonary artery and its branches. *Brit. Heart J.*, **26**, 832.
- Pool, P. E., Vogel, J. H., and Blount, S. G. (1962). Congenital unilateral absence of a pulmonary artery. *Amer. J. Cardiol.*, **10**, 706.
- Richter, K. (1963). *Die zentrale Lungenschlagader im Röntgenbild*. Akademie-Verlag, Berlin.
- Rockoff, D., and Gilbert, J. (1965). Functional pulmonary atresia. *Amer. J. Roentgenol.*, **94**, 85.
- Sherrick, D. W., Kincaid, O. W., and DuShane, J. W. (1962). Agenesis of a main branch of the pulmonary artery. *Amer. J. Roentgenol.*, **87**, 917.
- Smart, J., and Pattinson, J. N. (1956). Congenital absence of left pulmonary artery. *Brit. med. J.*, **1**, 491.
- Stecken, A. (1964). Ein weiterer Beitrag zur röntgenologischen Nativdiagnostik der Truncus-pulmonalis-Region im linksseitlichen Schichtbild. (Aplasie der linken Pulmonalarterie, Transposition der grossen Gefässe, Truncus arteriosus communis persistens.) *Fortschr. Röntgenstr.*, **100**, 181.